Case Report

Tuberculous Hypertrophic Pachymeningitis presenting as visual blurring and headaches

Rukhsana Tariq, Rashid Ahmed
Advanced Radiology Clinic, Off M A Jinnah Road, Karachi.
Corresponding Author: Rukhsana Tariq. Email: drrukhsanatariq@hotmail.com

Abstract

Hypertrophic cranial pachymeningitis is a rare chronic fibrosing inflammatory disease characterised by localized or diffuse thickening of duramater, leptomeninges, and tentorium. The etiology is diverse and includes infectious, granulomatous and inflammatory disorders, collagen vascular disorders, carcinoma, lymphoma, meningioma en plaque, sarcoidosis, haemodialysis, mucopolysaccharidosis, intrathecal drug administration, and meningeal carcinomatosis diseases. Intracranial hypotension is also an important image mimicker. Most often patients present with complaint of headache, vomiting, cranial nerve palsy, ataxia, raised intracranial pressure and focal neurological deficit. Other signs and symptoms are inconstant and variable. The imaging features of hypertrophic cranial pachymeningitis include dural thickening, dural mass, sinus thrombosis, venous congestion with white matter changes. Extensive preoperative imaging studies usually are essential by Computerised Tomography (CT) or Magnetic Resonance Imaging (MRI). The radiological findings may be characteristic of hypertrophic cranial pachymeningitis, may not divulge the underlying etiology. Meningeal biopsy is essential for diagnosing the cause. We reviewed a case of a tuberculous hypertrophic cranial pachymeningitis.

Keywords: Hypertrophic, Cranial, Pachymeningitis, Dura.

Introduction

Hypertrophic meningitis is a rare disorder characterized by diffuse or focal, linear or nodular thickening of the dura, underlying pia and arachnoid mater and the

Table: Causes of Hypertrophic Pachymeningitis.

Infection
Mycobacterium tuberculosis
Fungal infection
Cysticercosis
Human T-cell lymphotrophic virus I
Pseudomonas
Lyme disease
Syphilis
Systemic autoimmune diseases and vasculitides
Wegener granulomatosis
Rheumatoid arthritis
Sarcoidosis
Sjogren syndrome
Takayasu and Temporal arteritis
Idiopathic cranial or spinal pachymeningitis
Intracranial hypotension
Spontaneous hypotension
Hypotension occurring after spinal fluid drainage
Malignancy
Dural carcinomatosis
Metastatic disease in adjacent skull
Meningioma
Post surgical and VP shunt

tentorium.[1,2] Numerous causes leading to these changes have been identified (Table). Hypeutrophic Meningitis is more common in women[1] in the sixth decade of life.[3]

We present a case of focal and nodular pachymeningitis surrounding left temporal lobe.

Case Report

A 46 year old lady presented to our clinic with a history of headache and blurred vision. Her physical examination was normal. She had no motor or sensory deficits. Magnetic Resonance Imaging (MRI) performed demonstrated an extra axial, dural based extensive rind of soft tissue nodular mass in the left parasellar region, surrounding a portion of the left temporal lobe. The mass extended from high frontal convexity to the left clinoid process. The lesion was isointense to the parenchyma on T1 Weighted Image (T1WI), and hypointense on T2 Weighted Image (T2W) and Fluid Attenuated Recovery (FLAIR) images. Post contrast demonstrated diffuse homogenous enhancement of the lesion. There was vasogenic edema involving the underlying left temporal lobe (Figure-1). The mass was reported as granulomatous pachymeningitis, most likely due to tuberculous. The patient was referred for biopsy with partial resection. Histopathological diagnosis confirmed granulomatous inflammation, with tuberculosis reported as a possibility. The patient was put on antituberculous therapy with a good clinical response. Follow up MRI at 6months after surgery demonstrated persistence of meningeal nodularity around the left temporal lobe at the level of the left parasellar and Sylvian cistern, a finding consistent with hypertrophic pachymeningitis.

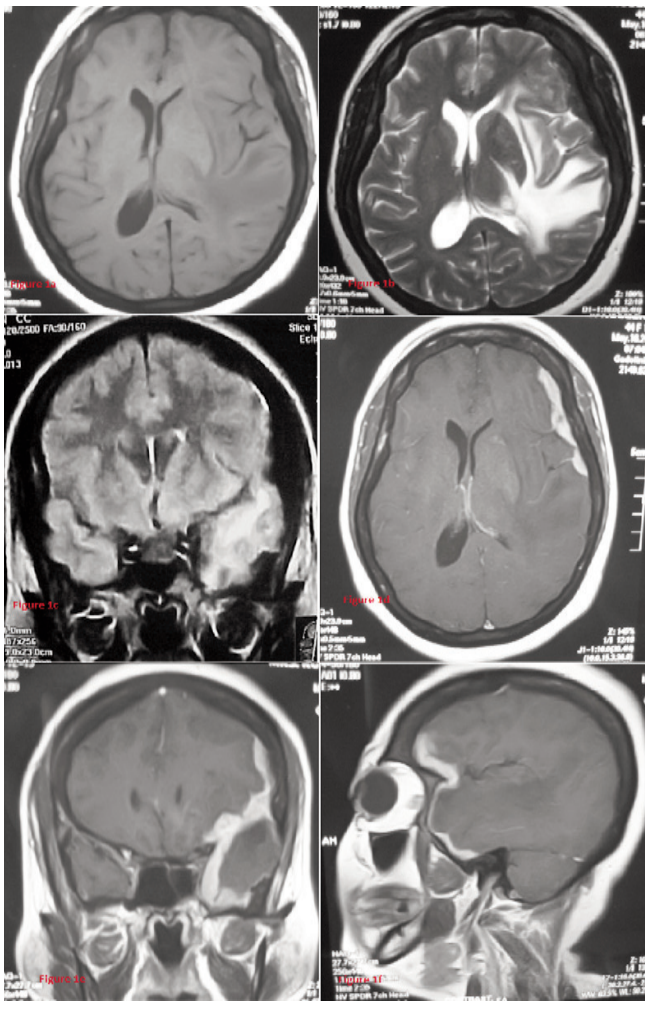

Figure-1: An extra axial, dural based, extensive rind of soft tissue nodular mass in the left parasellar region, surrounding a portion of the left temporal lobe. The mass extended from high frontal convexity to the left clinoid process. Vasogenic edema involving the underlying left temporal lobe and right subfalcine herniation is seen. (a)The lesion was isointense to the parenchyma on T1WI.(b & c) Hypointensity was seen on T2W and FLAIR images.(d,e,f) Post contrast axial, coronal and sagittal images demonstrate diffuse and homogenous enhancement of the lesion.

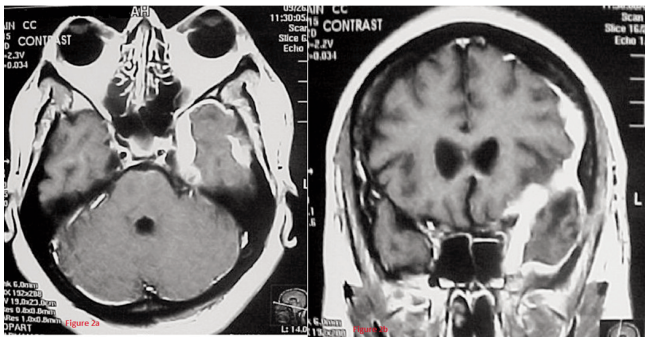

Figure-2: Postsurgical MRI after 6 months. Postcontrast axial (a) and coronal (b) images demonstrate residual enhancing meningeal nodularity, with decreased left temporal lobe edema and no midline shift.

Discussion

Hypertrophic pachymeningitis is an unusual rare inflammatory process involving the duramater, leptomeninges, and the tentorium. Multiple pathologic entities can produce pachymeningeal thickening. Hypertrophic pachymeningitis presents as nodular or linear thickening with intense meningeal enhancement.

These changes can be seen in infections, connective tissue disorders including Takayasu,[4] malignancies, post surgical, after placement of VP shunt, in intracranial hypotension[1,5] and can also be idiopathic, and may resemble neurosarcoid.[6] Clinically, chronic headaches, resembling migraine with or without other neurologic symptoms is the usual presentation.[1,2,7] Cranial nerve palsies, fits, cerebellar symptoms and opthalmological features including blindness, visual field loss, optic neuropathy, and raised intracranial pressure with papilledema have been reported. Headaches are attributed to focal meningeal irritation or secondary to localized arachnoiditis. Neurological

manifestations appear secondary to cranial nerve compression at the skull base due to meningeal hypertrophy resulting from chronic inflammatory process. Other complications including venous sinus thrombosis,[5] obstructive hydrocephalus,[5] cerebral oedema, Tolosa-Hunt syndrome, cranial neuropathies[1,7,8] have been noted. Besides, autoimmune diseases like Reidel thyroiditis, sclerosing cholangitis, multifocal fibrosclerosis and diabetes inspidus have been reported.[5]

Neuroimaging is essential for the preoperative diagnosis. CT reveals diffuse thickening and enhancement of the duramater.[2,5] Magnetic Resonance (MR) is advantageous to CT. On T1WI, smooth or nodular dural thickening with isointensity or hypointensity is seen. T2WI and FLAIR demonstrate hypointensity. Post Gadolinium shows avid and homogeneous enhancement .The signal characteristics are attributed to the fibrosis and necrosis of the duramater.[5,7] Peripheral hyperintensity on T2WI is due to active inflammation or increased vascularity of the duramater and underlying parenchyma.[5] The lesion is an avascular mass on angiography.[5] The treatment is intended to check the inflammation, responsible for morbidity associated with the disease. Treatment options include systemic corticosteroids, azathioprine and cyclophosphamide.[1,2,5,6,8] Immunomodulating agents[5] have been used in resistant cases. The clinical outcome ranges from complete resolution to progressive worsening of the disease leading to steroid dependence or surgical debulking. Follow up by MR to assess treatment efficacy is controversial as clinical improvement and imaging findings often do not correlate.[2,3,5]

In our case, the patient improved much more clinically on Anti Tuberculous Therapy (ATT) as compared to the radiolological improvement. Follow-up MR imaging performed 2 months after surgery revealed postsurgical changes in the left frontal lobe, resolution of midline shift, with no significant decrease in the axial dimension of the nodularity. MR imaging performed 6 months after surgery demonstrated persistence of meningeal nodularity, around the left temporal lobe at the level of the left parasellar and Sylvian cistern, a finding consistent with hypertrophic pachymeningitis.

Conclusion

Hypertrophic pachymeningitis is a rare disorder with many causative factors. The clinical outcome may range from complete resolution to progressive deterioration of the disease with the need for surgical debulking or steroid dependence. Imaging offers assessment of treatment efficiency that may remain contentious as clinical progress and imaging features often do not correlate, as was seen in our case. The case was unique on account of lack of clinical features and lab data for infection, with imaging manifestation supporting meningioma and neurosarcoidosis.

References

1. KupersmithMJ, Martin V, Heller G, Shah A, Mitnick HJ. Idiopathic hypertrophic pachymeningitis. Neurology 2004; 62: 686-94.
2. WangYJ, Fuh JL, Lirng JF, Lu SR, Wang SJ. Headache profile in patients with hypertrophic cranial pachymeningitis. Headache 2004; 44: 916-23.
3. KleiterI, Hans VH, Schuierer G, Marienhagen J, Han P, Schutz H, et al. Intraventricular cytarabine in a case of idiopathic hypertrophic pachymeningitis. J Neurol Neurosurg Psychiatry 2004; 75: 1346-8.
4. Wattamwar PR, Doshi SA, Thomas B, Nair M, Kuruvilla A. Hypertrophic Pachymeninngitis in a patient with Takayashu areteritis: One more association? Ann Indian Acad Neurol 2012; 15: 56-9.
5. Kazem IA, Robinette NL, Roosen N, Schaldenbrand MF, Kin JK. Best cases from the AFIP: Idiopathic tumefactive hypertrophic pachymeningitis. Radiographics 2005; 25: 1075-80.
6. Panos GC, Duarte GM, Pooia F. Idiopathic hypertrophic pachymeningitis mimicking neurosarcoidosis. Clinical Neurology and Neurosurgery 2012; 114: 176-8.
7. Riku S, Kato S. Idiopathic hypertrophic pachymeningitis. Neuropathology 2003; 23: 335-44.
8. Hatano N, Behari S, Nagatani T, Kimura M, Ooka K, Saito K, et al. Idiopathic hypertrophic cranial pachymeningitis: clinicoradiological spectrum and therapeutic options. Neurosurgery 1999; 45: 1336-43.